Essential Iris Atrophy

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Synonyms of Essential Iris Atrophy:

ICE syndrome, essential iris atrophy type iridocorneal endothelial (ICE) syndrome, essential iris atrophy progressive essential iris atrophy

General Discussion :

Essential iris atrophy is a very rare, progressive disorder of the eye characterized by a pupil that is out of place and/or distorted areas of degeneration on the iris (atrophy), and/or holes in the iris. This disorder most frequently affects only one eye (unilateral) and develops slowly over time. Attachment of portions of the iris to the cornea (peripheral anterior synechiae) and subsequent closure of the drainage angle may lead to secondary glaucoma and vision loss.

Essential iris atrophy is one of three iridocorneal endothelial (ICE) syndromes, each of which usually affects one eye of young to middle-aged men and women. The ICE syndromes (essential iris atrophy, Chandler syndrome, and Cogan-Reese syndrome) are distinct from one another. However, these disorders all affect the eye and some of their symptoms overlap, making it difficult to distinguish between them.

Signs & Symptoms :

Major symptoms of essential iris atrophy may include a displaced and/or distorted pupil, patchy areas of degeneration (atrophy) on the iris, and/or holes in the iris. The edge of the pupil may turn outward (ectropion uveae). The onset of this disorder is gradual, and the changes in the shape and placement of the pupil are usually noticed before any change in vision occurs. Degeneration and holes in the iris may develop over a period of several years.

Other features of essential iris atrophy may include the attachment of portions of the iris to the cornea (peripheral anterior synechiae), swelling of the cornea (corneal edema), and/or abnormalities in the cells lining the cornea (corneal endothelium). These changes may lead to increased pressure in the eye (glaucoma) and vision loss.

Causes :

The cause of essential iris atrophy or any other of the iridocorneal endothelial syndromes is not known. They are thought to be the result of the same mechanism. The primary defect is believed to be a cellular membrane secreted by the abnormal endothelial cells. This membrane covers the iris and the drainage angle of the eye. The contraction of this membrane leads to pupillary changes and formation of peripheral anterior synechiae with resultant angle closure glaucoma.

Other researchers suspect that inflammation or chronic infection may be the cause of the disease. There is a hypothesis that ICE syndromes stem from an in-vitro herpes infection localized in the endothelial layer. According to this theory, one eye is infected first and the second eye develops immunity before it can be affected.

Affected Populations :

Essential iris atrophy is a very rare disorder that predominantly affects females in the middle adult years. The prevalence is not known.

Diagnosis :

Symptoms of the following disorders can be similar to those of essential iris atrophy. Comparisons may be useful for a differential diagnosis:

Chandler's syndrome (CS) is a rare eye disorder in which the endothelium, the single layer of cells lining the inner surface of the cornea, proliferates causing corneal edema, distortion of the iris, and unusually high pressure in the eye (glaucoma).

The spectrum is an acquired, unilateral disorder, which typically occurs in early to middle adulthood and predominantly affects women. Chandler's syndrome is the most commonly encountered clinical variant of this spectrum.

Cogan-Reese syndrome is an extremely rare disorder characterized by loss of iris tissue and the development of small wart-like growths on the iris. Increased pressure within the eye (glaucoma) and corneal swelling (edema) are also evident. This disorder differs from Cogan corneal dystrophy which is inherited as an autosomal dominant disorder. The displacement and/or distortion of the pupil characteristic of essential iris atrophy does not occur in Cogan corneal dystrophy.

Axenfeld's anomaly is characterized by attachment of portions of the iris to the cornea (peripheral anterior synechiae). Axenfeld's anomaly is considered to be an inherited, developmental defect, while the iridocorneal syndromes (Cogan-Reese syndrome, Chandler's syndrome, and essential iris atrophy) are thought to be acquired disorders.

There is some confusion in the medical literature as to whether Axenfeld's and Rieger's anomalies are separate disorders or whether they occur together in what is called the Axenfeld-Rieger (A-R) syndrome. Of note, A-R syndrome is a bilateral condition, whereas ICE syndrome is usually unilateral.

Rieger's anomaly is characterized by attachment of portions of the iris to the cornea, a distorted pupil, clouding of the edges of the cornea (peripheral corneal opacification), displacement of iris tissue (hypoplasia), and/or secondary glaucoma.

When Rieger's anomaly occurs in association with dental abnormalities (i.e., a decrease in the number of teeth, small teeth, or anodontia) and facial malformations (i.e., displacement of the jaw, flattening of the midface, a receding upper lip and prominent lower lip) it is referred to as Rieger's syndrome. Rieger anomaly is considered to be an inherited, developmental defect.

Standard Therapies :

Secondary Glaucoma and Treatment:

Glaucoma may occur as a secondary disorder to essential iris atrophy. The mechanism of glaucoma in ICE syndrome (all three variants) is believed to be related to a cellular membrane secreted by the abnormal endothelial cells. This membrane covers the trabecular meshwork of the drainage angle, thereby obstructing aqueous outflow facility and elevating intraocular pressure. In the early stages, the angle may appear open clinically although it is covered by this transparent membrane. Over time, contraction of this membrane leads to peripheral anterior synechiae and secondary angle closure glaucoma.

Treatment of essential iris atrophy usually involves the use of drops in the eyes to control the glaucoma and swelling (edema). Mild cases or corneal edema are often managed with soft contact lenses and hypertonic saline solutions. In advanced cases penetrating or endothelial keratoplasty may be required, although the failure rate is high with need for repeat corneal grafts. In some individuals, the corneal edema may be improved with reduction in intraocular pressure. Medical therapy for glaucoma is usually initiated with aqueous suppressants, including beta blockers, alpha-2 agonists and carbonic anhydrase inhibitors.

Prostaglandin analogues may be helpful in some cases. Surgical intervention for glaucoma is eventually required in a high percentage of patients with ICE syndrome. The most commonly performed procedure is trabeculectomy, with variable success rates. Glaucoma drainage devices have shown favorable outcomes in a small number of patients, but further studies are warranted to validate these results in a large series. Laser surgery is rarely effective.

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